CASE REPORT

Basedow paraplegia: A possible misnomer

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Thyrotoxic myopathy frequently occurs in clinical practice; however, the association of hyperthyroidism with a flaccid, areflexic paraplegia, so-called Basedow paraplegia, appears to represent a controversial and doubtful entity.

An 18-year-old female with undiagnosed and untreated Graves' disease presented with acute onset of global weakness predominantly in the lower limbs, but also affecting the upper limbs. The weakness was accompanied by hypotonia and areflexia. Clinically, the patient had a goitre and signs of thyroid ocular disease. Laboratory testing confirmed the presence of hyperthyroidism, and thyroid-stimulating hormone receptor antibodies were positive. The cerebrospinal fluid protein level was raised. The electroneuronographic and needle examinations were compatible with a clear denervation process, such as acute motor axonal neuropathy, a variant of Guillain-Barré syndrome. Intravenous immunoglobulin therapy, carbimazole and propranolol were administered.

The occurrence of hyperthyroidism with a flaccid, areflexic paraplegia appears to represent more of a fortuitous than a causative association. It is important to consider and treat other causes, such as acute idiopathic polyneuritis.

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Common neurological complications of untreated Graves' disease include cognitive dysfunction, tremor, ophthalmopathy, myopathy and polyneuropathy. Myasthenia gravis and seizures are uncommon associations, while thyrotoxic periodic paralysis, stroke and chorea occur only rarely.[1]

We present a patient with Graves' disease and the acute motor axonal neuropathy (AMAN) variant of Guillain-Barré syndrome, which masqueraded as so-called Basedow paraplegia.[1-5] Had the diagnosis of Basedow paraplegia been adhered to, the patient would have been denied the opportunity of receiving gammaglobulin therapy.

Case report

An 18-year-old female was admitted in November 2014 with acute onset of severe global leg and arm weakness that had started 4 days before her admission. There was a background history (over the previous 11 months) of proptosis, dyspnoea on exertion, palpitations, irritability and forgetfulness. General examination revealed tachycardia and a symmetrical diffusely enlarged goitre. Proptosis and lid lag were also present. Neurological examination revealed motor weakness, with her legs more affected than her arms. There was global hypotonia and deep tendon areflexia, with sparing of all sensory modalities. Blood tests confirmed the clinical suspicion of hyperthyroidism. Serum potassium levels were repeatedly normal (Table 1), urine porphobilinogen screening was negative and antiganglioside antibodies were absent.

Table 1. Laboratory results

	On admission	1 week after admission	2 weeks after admission
TSH (mIU/L) (0.48 - 4.26)	0.08		
Free thyroxine (pmol/L) (7.6 - 16.1)	55	30.4	27.5
Potassium (mmol/L) (3.5 - 5.1)	4.9	4.0	
TSH receptor antibody (U/L) (<1.75)	36.26		
Cerebrospinal fluid protein (g/L) (0.15 - 0.45)	0.86		

TSH = thyroid-stimulating hormone.

Table 2. Electroneuronographic study, 11 November 2014

		Motor nerve responses		Sensory nerve responses		
Nerve	Muscle	CMAP amplitude	Normal values	Peak latency	SNAP amplitude	Normal value
R median	APB	2.6 mV	>4.0 mV	Palmar	50.7 µV	>50 µV
R ulnar	ADM	530 µV	>6 mV	Palmar	34.5 µV	>15 µV
R peroneal	EDB	127 µV	>2 mV			
R tibial	AH	1.7 mV	>4 mV			
R sural				Point B	24.0 µV	>6 µV

R = right; CMAP = compound muscle action potential; SNAP = sensory nerve action potential; APB = abductor pollicis brevis; ADM = abductor digiti minimi; EDB = extensor digitorum brevis; AH = abductor hallices. The R median, ulnar, peroneal and tibial nerve latencies were 2.8 ms, 2.4 ms, 3.0 ms and 3.5 ms, respectively (within normal limits), and their conduction velocities 54.4 m/s, 70.3 m/s, 43.4 m/s and 55.3 m/s, respectively, were also normal. The sensory peak latencies of the R median (palmar), R ulnar (palmar) and R sural (point B) nerves were 1.9 ms, 2.0 ms and 3.3 ms, respectively (within normal limits), and their conduction velocities 56.3 m/s, 51.3 m/s and 51.5 m/s, respectively (within normal limits).

Table 3. Electroneuronographic study, 25 November 2014

	Motor nerve responses			Sensory nerve responses		
Nerve	Muscle	CMAP amplitude	Normal values	Peak latency	SNAP amplitude	Normal value
L median	APB	2.2 mV	>4.0 mV	Palmar	52.3 µV	>50 µV
L ulnar	ADM	339.0 µV	>6 mV	Palmar	17.9 µV	>15 µV
L peroneal	EDB	195.0 µV	>2 mV			
R peroneal	EDB	92 µV	>2 mV			
L tibial	AH	No response	>4 mV			
R tibial	AH	853 µV	>4 mV			
L sural				Point B	14.1 µV	>6 µV

L = left; R = right; CMAP = compound muscle action potential; SNAP = sensory nerve action potential. The L median, L ulnar, L peroneal, R peroneal and R tibial nerve distal latencies were 2.6 ms, 2.6 ms, 4.4 ms, 4.9 ms and 3.8 ms, respectively (within normal limits), and their conduction velocities 46.8 m/s, 56.6 m/s, 43.2 m/s, 50.0 m/s and 52.2 m/s, respectively, were also 'essentially' normal. The sensory peak latencies of the R median (palmar), R ulnar (palmar) and R sural (point B) nerves were 2.0 ms, 2.2 ms and 3.8 ms, respectively (within normal limits), and their conduction velocities 54.6 m/s, 50.6 m/s and 46.1 m/s, respectively, were also 'essentially' normal.

The electroneuronographic studies of 11 and 25 November 2014 showed significantly decreased compound muscle action potential (CMAP) amplitudes and preserved sensory nerve action potential (SNAP) amplitudes, with normal distal latencies and conduction velocities, favouring a diagnosis of an AMAN variant of the Guillain-Barré syndrome (Tables 2 and 3). Needle examination of the tibialis anterior muscle on 25 November 2014 showed the presence of fibrillation potentials and clear neurogenic polyphasic motor units.

A 5-day course of intravenous immunoglobulins, 24 g/day, was administered. Carbimazole was prescribed at 20 mg 8-hourly, and on the development of a skin reaction the dose was decreased to 10 mg 8-hourly. Propranonol was administered at a dose of 20 mg 6-hourly. The thyrotoxicosis gradually improved, but the patient's neurological condition had only marginally improved at the time of discharge.

Discussion

Joffroy coined the term Basedow paraplegia in 1894, after Charcot had referred to 'paraplegia like weakness' in severe hyperthyroidism in 1889.[3] Joffroy went on to describe the concept as follows: 'A flaccid paraplegia with absent reflexes, minimal or no sensory disturbance, and absent sphincter disturbances,'[3] which Pandit in 1998 commented 'could very well fit the description of acute post infective polyneuritis'.[3] Feibel and Campa[2] used the term 'thyrotoxic neuropathy (Basedow paraplegia)' in 1976 and Pandit,[3] in 1998, published a report 'Acute thyrotoxic neuropathy – Basedow's paraplegia revisited,' with the suggestion of an implicit causal relationship between the hyperthyroidism and the neuropathy. However, on closer scrutiny this association may well be fortuitous.

Descriptions of Basedow paraplegia[1-5] appear to conform to the development of an 'acute flaccid paraplegia with absent reflexes' against the background of hyperthyroidism. It is, however, possible that this clinical presentation may reflect the occurrence of an acute idiopathic polyneuritis,[6] possibly associated with an underlying predisposition to autoimmune diseases.[1]

The association between hyperthyroidism and acute flaccid areflexic neuropathy receives little credence in the following well-known clinical textbooks: Dyck and Thomas' *Peripheral Neuropathy*[7] comments on its uncertain association and the difficulty to distinguish it from acute idiopathic polyneuritis; *Bradley's Neurology in Clinical Practice*[8] refers to its association as fortuitous; and *Williams's Textbook of Endocrinology*[9] and *Harrison's Internal Medicine*[10] do not even mention the association.

Conclusion

It is important to consider the occurrence of other treatable causes of motor paralysis in patients with Graves' disease, such as acute idiopathic polyneuritis presenting with a rapid onset of flaccid paralysis. The entity of Basedow paraplegia as a diagnosis, *per se*, was found to be misleading.

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